International Journal of Otorhinolaryngology and Head and Neck Surgery
Gopal A et al. Int J Otorhinolaryngol Head Neck Surg. 2022 Dec;8(12):1012-1016
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

DOI: https://dx.doi.org/10.18203/issn.2454-5929.ijohns20223055

Case Report

Plasma cell mucositis masquerading as squamous cell carcinoma-a surgical dilemma: a rare case report and review of literature

Ashish Gopal[1], Maya Singh[1]*, Ishwar Singh[1], Sharmana Mandal[2]

[1]Department of Otorhinolaryngology and Head and Neck Surgery, Maulana Azad Medical College, New Delhi, India
[2]Department of Pathology, Maulana Azad Medical College, New Delhi, India

Received: 08 September 2022
Accepted: 20 October 2022

***Correspondence:**
Dr. Maya Singh,
E-mail: singhmaya183@gmail.com

ABSTRACT

Plasma cell mucositis is a rare, idiopathic, multifocal, benign proliferative disorder affecting upper aerodigestive tract. It is a very rare condition and less than 50 cases reported in literature. A 62 years old female patient reported with complaints of oral discomfort, superficial erosive ulceration on the tongue, and burning sensation on eating food for last 2 years. Initially, Incisional biopsy of the involved mucosal area confirmed the diagnosis of well differentiated squamous cell carcinoma for which hemi-glossectomy was done. However histopathological examination submitted revealed a hyperplastic epithelium with a dense infiltrate of mature polyclonal plasma cells in the superficial layer of lamina propria, for which plasma cell mucositis was made a diagnosis. Plasma cell mucositis is a diagnosis of exclusion, to be differentiated from other infective, reactive, autoimmune, allergic and neoplastic disorders with plasma cell infiltrates. Management includes topical, intralesional, systemic corticosteroids and debulking procedures. Regular follow-up is essential due to the possibility of its clinical persistence and chances of malignant transformation.

Keywords: Plasma cell mucositis, Squamous cell carcinoma, Plasmacytosis

INTRODUCTION

Plasma cell mucositis is an unusual plasma cell proliferative disorder of the upper aerodigestive tract.[1]

Plasma cell infiltrates in connective tissue can be seen in malignant, reactive, autoimmune, infectious and idiopathic conditions. In 1952, Zoon first described plasma cell infiltrates in the glans penis, which he called as balanitis plasma cellularis. This was reported by others as well, and came to be known as Zoon's balanitis.[2]

Plasma cell mucositis is an extremely rare condition, with less than 50 cases reported so far. It is a benign inflammatory condition distinguished by dense plasma cell infiltrate in the mucosa.[3] It was previously reported as idiopathic plasmacytosis, plasma cell orifical mucositis, oral papillary plasmacytosis.[4-6]

It is a benign polyclonal plasma cell proliferative disorder of mucous membrane with an unknown aetiology. There is a slight male predominance of 1:2.1 and the average age of presentation is 56.6 years. The clinical appearances of plasma cell mucositis is varied, but typically it presents as a florid erythematous oral mucosa with nodular, cobblestone, papillomatous, granular, velvety surface changes.[4]

Plasma cell mucositis is a diagnosis of exclusion, requiring extensive investigations and multidisciplinary evaluation as the clinical and histological features of plasma cell mucositis resemble many common benign and neoplastic conditions of oral cavity.

There is just one case reported in literature where a Squamous Cell Carcinoma arose from a mucosal plasmacytosis and the aetiology is based upon the

immunosuppressive drugs given for plasma cell mucositis.[5] No further review of literature is available for such a case.

A rare case of plasma cell mucositis, presenting as squamous cell carcinoma on incisional biopsy in an elderly female patient causing stomato-dynia and dysphagia is presented. The challenges encountered while making a diagnosis and management is also described.

CASE REPORT

A 62 years old female patient, presented to the department of otorhinolaryngology and head and neck surgery, Lok Nayak hospital with complaints of oral discomfort, superficial erosive ulceration and burning sensation in her mouth on eating food, local dysgeusia, pain over the tip of tongue and both sides of tongue for past 2 years. (Figure 1).

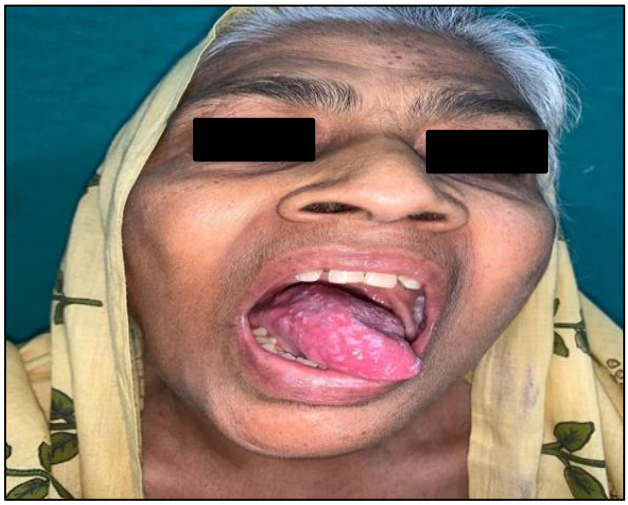

Figure 1: Preoperative lesions of the tongue.

The patient reported that burning sensation was gradually progressive, which was more on the left side compared to the right side of the tongue. She now complained of sore throat and difficulty in swallowing solids since past few months forcing her to have semisolid and liquid bland diet.

She did not have any constitutional symptoms like fever, loss of appetite/weight, fatigue and arthralgia.

She denied any smoking, tobacco or alcohol consumption and did not have any systemic illness. There was no history of allergies or family history of similar conditions.

There was no past history of hypertension, diabetes mellitus. She had past history of laparoscopic cholecystectomy under general anaesthesia in 2011.

Examination revealed intraorally reddish coloured superficial erosive ulceration present on both right and left lateral border, dorsum and tip of tongue, which was indurated. There was no evidence of any bullae, scarring, vesicles. The gingiva, palatal and oropharyngeal mucosa was erythematous with velvety shiny irregular surface. (Figure 2 A and B).

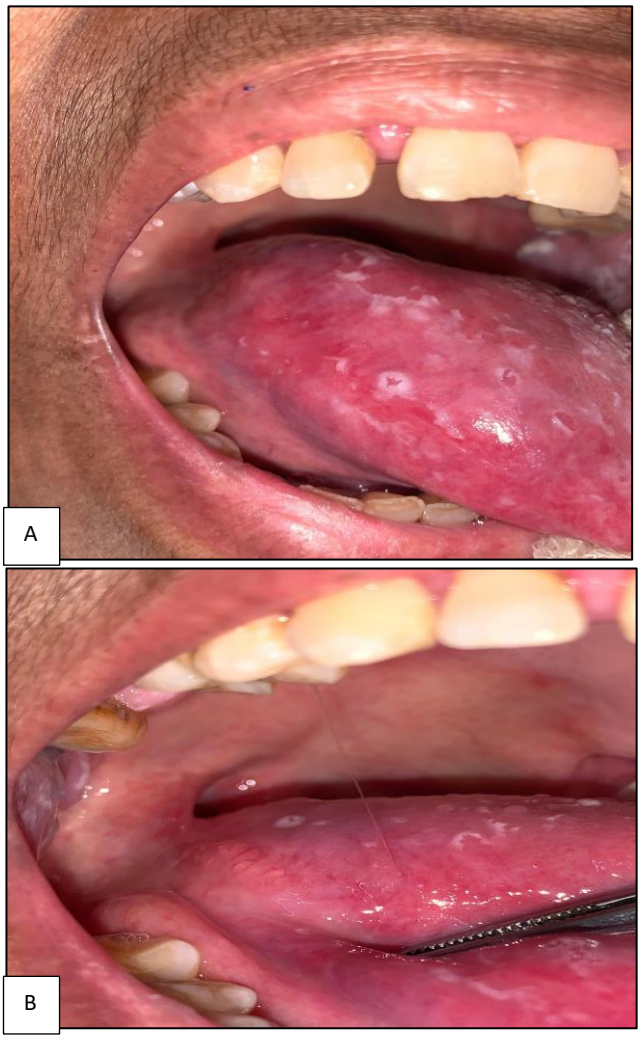

Figure 2 (A and B): Erosive lesions on right lateral border tongue.

External examination revealed normal facial morphology, and sub centimetric cervical lymph node swelling on the left side.

Based on the clinical findings, this condition could be due to an oral manifestation of other systemic illness or due to any local factors.

The patient was prescribed topical corticosteroids (Triamcinolone acetonide gel 0.1% for topical application 4 times daily, with chlorhexidine mouth wash). Lidocaine gel was prescribed for burning sensation before meals to prevent discomfort while having food. The patient was called for follow up weekly. After 2-month follow-up, there was mild relief in the burning sensation. However, there was no improvement in the erythematous, reddish, erosive lesions present on the tongue.

The patient was advised for an incisional biopsy under local anaesthesia from right and left lateral border of tongue.

After the patient provided written informed consent, the soft tissue from left and right lateral borders was biopsied. The biopsied specimen from left lateral half of tongue showed well differentiated squamous cell carcinoma, whereas the right half of tongue came out to be plasma cell mucositis.

Radiological imaging of head, neck stated that patient is a suspected case of carcinoma tongue, findings do not suggest any infiltrative mass lesion of tongue, the lesion is likely to involve the superficial cutaneous planes with no evidence of significant cervical lymph nodes are seen (Figure 3).

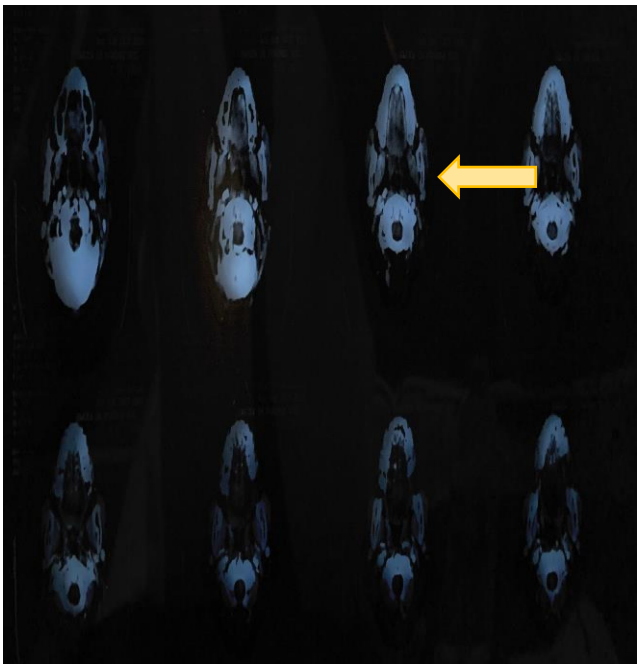

Figure 3: Contrast enhanced computed tomography of base of skull to diaphragm, showing infiltrative mass lesion of tongue, involving superficial cutaneous planes (arrows).

On the basis of clinical suspicion and histopathological analysis, left hemi glossectomy with supra-omohyoid neck dissection was done.

The histopathological examination from the left hemi-glossectomy specimen showed, focal ulceration of lining stratified squamous epithelium with ulcer bed formed by acute on chronic inflammatory granulation tissue. There is predominance of plasma cells with admixed neutrophils and lymphocytes. Focal mild to moderate fibrosis was seen along with the plasma cell infiltrate, extending into underlying muscles. The tumour cells showed a polyclonal expression of Kappa, lambda and expression of CD38, CD138, and negative for IgG, IgG4, CD34, CD20, CD3, PAX5. The impression of plasma cell mucositis was made and no evidence of squamous cell carcinoma was mentioned as reported earlier in the incisional biopsy report (Figure 4 and 5).

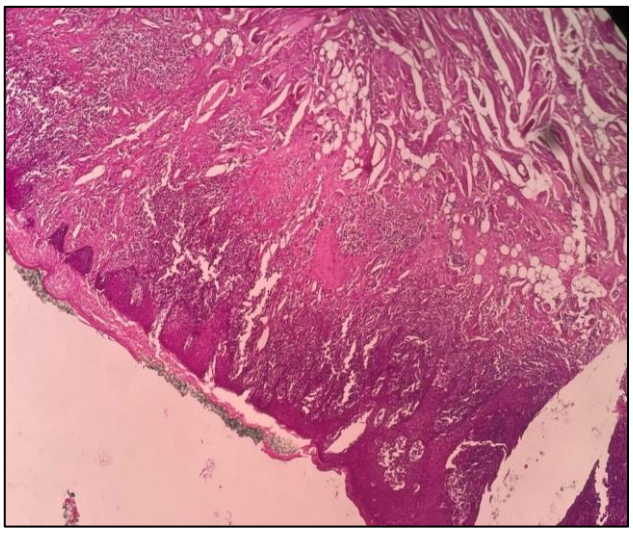

Figure 4: Lining of stratified squamous epithelium with focal ulceration, sub epithelium shows sheets of plasma cells (HEx109).

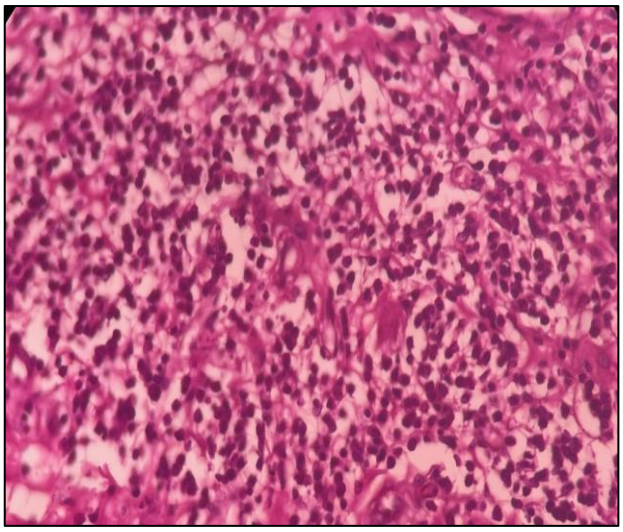

Figure 5: Sheets of plasma cells (HE x 400).

Therefore, the patient was advised to get a bone scan, serum electrophoresis, Bence jones proteins and haematological work up to rule out plasma cell dyscrasia.

Routine blood tests, including full blood count with white cell differential, renal, thyroid, liver function tests, in addition to chest x-ray, ultrasonography of neck were all normal.

Other laboratory tests to screen for systemic myeloma was performed. These included serum and urine protein electrophoresis, measurements of the erythrocyte sedimentation rate, and the levels of immunoglobulins

and urinary Bence-Jones proteins. All results were normal.

On the basis of above results, diagnosis of plasma cell mucositis was made.

The patient was then prescribed systemic corticosteroids prednisolone 60 mg once daily with topical application of antifungal mouth paint and topical corticosteroid on the tongue. Proton pump inhibitors with calcium supplements was prescribed along to prevent side effects of long-term corticosteroid therapy.

She was followed weekly and after 2 months follow up, the symptoms were relieved. Oral cavity examination, significant improvement in the clinical appearance of gingival, and superficial erosive ulceration on the other half of the tongue. The topical and systemic steroids was gradually tapered off over the next 6 weeks.

DISCUSSION

Plasma cell mucositis is a benign condition which is extremely rare, less than 50 cases reported so far.[6,7]

It can affect the oral cavity, as well as nasopharynx, oropharynx, larynx, hypopharynx, and esophagus.

Dysphagia, pharyngitis, stridor, oral pain, chronic cough, hoarseness, dyspnea, stridor are the presenting symptoms.[1]

Epithelial hyperplasia with acanthosis and spongiosis is some histological features that are reported. The epithelium shows psoriasiform or pseudo-epitheliomatous hyperplasia. Superficial lamina propria shows dense subepithelial plasmacytic infiltration. No atypia or prominent nucleoli is seen in mature plasma cells. Occasionally Russell bodies may be present. As a result of ulceration and secondary non- specific inflammation, there may be few polymorphonuclear leucocytes and lymphocytes with exocytosis and micro abscess formation. Polyclonal plasma cell infiltration with mixed population of kappa and lambda light chains and various heavy chains are seen in immune-peroxidase staining.[4,8]

In this study, focal ulceration of lining stratified squamous epithelium with ulcer bed formed by acute on chronic inflammatory granulation tissue was seen. There was predominance of plasma cells with admixed neutrophils and lymphocytes. Focal mild to moderate fibrosis was seen along with the plasma cell infiltrate, extending into underlying muscles. The tumour cells showed a polyclonal expression of Kappa, lambda and expression of CD38, CD138, and negative for IgG, IgG4, CD34, CD20, CD3, PAX5. There was no evidence of carcinoma, amyloid deposits or granulomas.

Since it is a very rare pathology, it is important to exclude other diseases with similar pathological features, such as extramedullary plasmacytoma, erosive lichen planus, mucous membrane pemphigoid, allergic gingivostomatitis, squamous cell carcinoma and fungal infections.[9,10]

A rare peripheral B cell neoplasm comprising 1% of all head and neck tumours is extramedullary plasmacytoma. It constitutes one of the three variants of plasma cell neoplasms, the other two being solitary bone plasmacytoma or (medullary plasmacytoma) and multiple myeloma.[11]

Extramedullary plasmacytoma shows an inclination towards head and neck region with 80% of cases occurring in upper aerodigestive tract.[12] Men are more commonly affected than women with a male: female ratio of 3;1. 55 years is the median age of presentation.[13]

Many times, plasma cell mucositis can enter into differential diagnosis with squamous cell carcinoma, in literature there is a case report where a squamous cell carcinoma arose from a mucosal plasmacytosis.[5,10] Hence, it is very important to examine the benign/neoplastic nature of plasma cell infiltrate, as the management and prognosis of plasma cell neoplasms are very different from benign conditions. When immunohistochemistry results are inconclusive, gene rearrangement studies can be done to find a diagnosis.

In this case, all the differential diagnosis were excluded based on investigations and a diagnosis of plasma cell mucositis was made.

The management of plasma cell mucositis is challenging, aimed at reducing the symptoms. The adverse side effects of topical and systemic steroids limit its prolonged use. Methotrexate, dapsone, tacrolimus, cyclosporine, mycophenolate mofetil, azathioprine infliximab, adalimumab and golimumab are various immunosuppressive agents that have been used with varying success. When using potent immunosuppressive agents, caution should be exercised for this benign condition where risk factors for malignancy exist.[5]

However, the use of platelet rich fibrin injections with cortisone therapy are reported in some cases, in literature.[14]

Use of low dose radiotherapy, and systemic chemotherapy (vincristine, prednisolone, cyclophosphamide) have been shown in severe disease. Excision by carbon dioxide lasers, or debulking procedures like surgical excision, cryotherapy, electrocoagulation has provided temporary relief followed by recurrence.[1,4,5,9]

In cases of subglottic stricture or airway compromise, surgical intervention is required, with tracheostomy being done in two reported cases.[1,15]

When considering a rare diagnosis such as plasma cell mucositis based on clinical pathological correlation, it is important to report and recognise such cases for awareness of its management as squamous cell carcinoma can enter into a differential diagnosis of plasma cell mucositis.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Ferreiro JA, Egorshin EV, Olsen KD, Banks PM, Weiland LH. Mucous membrane plasmacytosis of the upper aerodigestive tract. A clinicopathologic study. Am J Surg Pathol. 1994;18(10):1048-53.
2. Zoon JJ. Chronic benign circumscript plasmocytic balanoposthitis. Dermatologica. 1952;105:1-7.
3. Puvanendran M, Lieder A, Issing W. Plasma cell mucositis of oro-and hypopharynx: a case report. Case Rep Otolaryngol. 2012;2012.
4. Solomon LW, Wein RO, Rosenwald I, Laver N. Plasma cell mucositis of the oral cavity: Report of a case and review of the literature. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2008;106:853-60.
5. Pepper T, Shekar K, Singh M, Brennan PA. Squamous cell carcinoma arising in mucosal plasmacytosis. Br J Oral Maxillofac Surg. 2010;48:208-10.
6. White JW, Jr, Olsen KD, Banks PM. Plasma cell orificial mucositis. Report of a case and review of the literature. Arch Dermatol. 1986;122:1321-4.
7. Aiba S, Tagami H. Immunoglobulin-producing cells in plasma cell orificial mucositis. J Cutan Pathol. 1989;16(4):207-10.
8. Bharti R, Smith DR. Mucous membrane plasmacytosis: a case report and review of the literature. Dermatol Online J. 2003;9(5):15-5.
9. Smith ME, Crighton AJ, Chisholm DM, Mountain RE. Plasma cell mucositis: a review and case report. J Oral Pathol Med. 1999;28(4):183-6.
10. Khan NA, McKerrow WS, Palmer TJ. Mucous membrane plasmacytosis of the upper aerodigestive tract. A case report with effective treatment. J Laryngol Otol. 1997;111(3):293-5.
11. Iseri M, Ozturk M, Ulubil SA. Synchronous presentation of extramedullary plasmacytoma in the nasopharynx and the larynx. Ear Nose Throat J. 2009;88(11):E9-12.
12. Galieni P, Cavo M, Pulsoni A, Avvisati G, Bigazzi C, Neri S et al. Clinical outcome of extramedullary plasmacytoma. Haematologica. 2000;85(1):47-51.
13. Straetmans J, Stokroos R. Extramedullary plasmacytomas in the head and neck region. Eur Arch Oto-Rhino-Laryngol. 2008;265(11):1417-23.
14. Gasparro R, Adamo D, Masucci M, Sammartino G, Mignogna MD. Use of injectable platelet-rich fibrin in the treatment of plasma cell mucositis of the oral cavity refractory to corticosteroid therapy: A case report. Dermatolo Therap. 2019;32(5):e13062-2
15. Galvin S, Bowe C, Regan EM, Conlon N, Flint SR, Healy CM. Circumorificial plasmacytosis/plasma cell orificial mucositis: a case series and a review of the literature. Oral Surg Oral Med Oral Pathol Oral Radiol. 2016;122(3):e77-81.

Cite this article as: Gopal A, Singh M, Singh I, Mandal S. Plasma cell mucositis masquerading as squamous cell carcinoma-a surgical dilemma: a rare case report and review of literature. Int J Otorhinolaryngol Head Neck Surg 2022;8:1012-6.